Visual Field Defects in Deformational Posterior Plagiocephaly

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Purpose: We sought to determine whether visual field abnormalities occur in infants with deformational posterior plagiocephaly and to assess whether there is a relationship between the severity and laterality of visual field abnormalities with the severity and laterality of skull deformity. ***Methods:*** A retrospective chart review was performed on 40 consecutive infants with deformational posterior plagiocephaly. Each was tested with standardized binocular arc perimetry in the horizontal plane. Sixteen patients also had 3-dimensional computed tomography of the skull. Hemifield asymmetry of $\geq$20 degrees and/or a decrease in hemifield values by $\geq$20 degrees from established normal patients was considered abnormal. Visual field data from study patients was plotted against previously published normative data. Graphs comparing visual field defects and laterality to cranial asymmetry also were generated. ***Results:*** Thirty-five percent of infants with deformational posterior plagiocephaly had constriction of one or both hemifields by at least 20 degrees from established normal patients. Hemifield asymmetry of 20 degrees or more was found in 17.5% of infants tested. There was a significant difference in the worse hemifield values measured in each patient and the standard visual fields obtained from normative data ($P = 0.036$). There was no correlation between the laterality of the visual fields to the laterality of the defects. A correlation between severity of hemifield constriction and % asymmetry on computed tomography was noted ($P = 0.209$). ***Conclusions:*** Deformational posterior plagiocephaly may affect visual field development but neither the laterality nor the severity of skull deformity is predictive of the severity of visual field defects. (J AAPOS 2005; 9:274-278)

Posterior plagiocephaly is an abnormality of the infant skull resulting in unilateral flattening of the occiput and ipsilateral frontal protrusion (Figures 1 A and B).[1] Posterior plagiocephaly can be the result of lambdoid synostosis in rare cases, but it is more commonly caused by a deformational, nonsynostotic mechanism.[1-5] In the 1970s, the incidence was reported to be approximately 1 in 300,[4] but more recent literature estimates a rate between 8% and 12% in infancy, increasing to as much as 47% at 1 year, [6,7] although a true incidence value is really unknown.[8] Although most studies demonstrate a higher frequency in boys[3,7,9] a few studies have shown girls to be more affected.[5]

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According to Rekate, some degree of deformational posterior plagiocephaly may be morphometrically present in 14% of adults and often goes unrecognized.[8] Most authors note a left-sided predominance as well.[5]

The etiology of posterior plagiocephaly is likely multifactorial, but predisposing factors have been noted, including positioning preference, torticollis, prematurity, and developmental delay.[2] There has been an increase in the incidence of deformational posterior plagiocephaly correlating with the 1992 recommendations by the American Academy of Pediatrics to change infants to a supine or side sleeping position to decrease the risk of sudden infant death syndrome (SIDS).[10,11] According to the American Academy of Pediatrics Task Force on Infant Positioning and SIDS, the number of infants who slept supine more than doubled (30–76%) from 1992 to 1996.[11] Referrals to the North Carolina Center for Craniofacial Deformities rose more than 4-fold from 1990 to 1994, and the incidence of plagiocephaly increased by a factor of 10.[10]

Deformational posterior plagiocephaly has been anecdotally associated with visual field and other ophthalmologic and neurologic deficits, such as astigmatism and strabismus.[12,13] If detected in the first year of life, it is treatable with noninvasive means, such as change in positioning or helmet therapy (Figure 2).[3] Therefore, it is important to determine

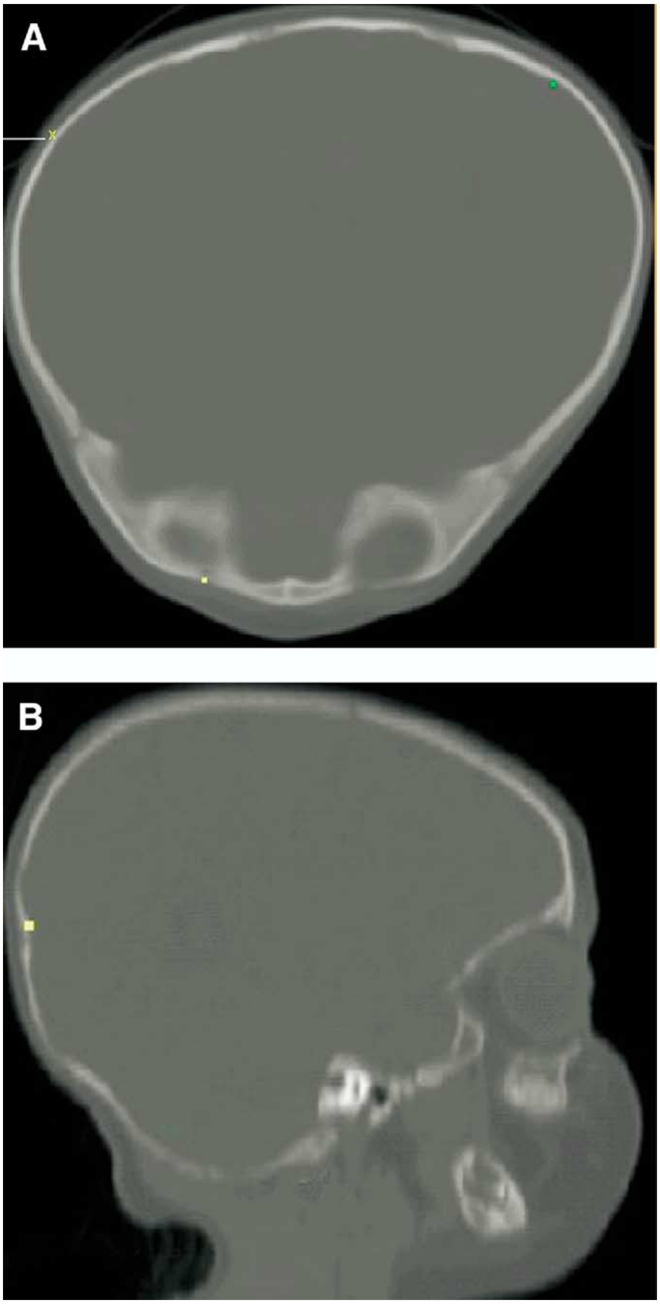

FIG 1. Head CT (bone windows) showing posterior plagiocephaly. A, Axial view. B, Sagittal view.

whether any quantifiable, clinically relevant neurological deficits are associated with posterior plagiocephaly, and if treatment improves these deficits. Visual field testing has been suggested as a means of detecting abnormalities in cortical pathway maturation in infants.[14,15] Using the normative data obtained by a previous study[14] and attempting to duplicate the testing method and environment, visual field testing was performed in a cohort of patients with posterior plagiocephaly to document the presence and severity of visual field abnormalities in this group.

FIG 2. Photograph of an infant with deformational posterior plagiocephaly wearing an orthotic helmet.

METHODS

After approval from the University of Oklahoma Health Sciences Center Institutional Review Board, a retrospective chart review of 40 consecutive infants, ages 19-53 weeks, with a diagnosis of deformational posterior plagiocephaly and otherwise-normal ophthalmologic examination was performed during a 3-year period. Patients were included if they had no prior helmet therapy, were able to cooperate for visual field testing, had normal Teller acuities (Vistech Consultants, Inc., Seattle, WA), and had no strabismus, amblyopia, or retinal or optic nerve abnormalities; on these bases, they were considered likely to have normal binocular visual function for age. Standardized binocular arc perimetry (Ferree-Rand Simplified Perimeter, Bausch and Lomb, Rochester, NY) in the horizontal plane, as previously described,[12] was performed on each child by 2 separate examiners masked to each other, measuring the temporal right and left hemifields using a preferential looking technique with a target subtending an angle of 6 degrees. The visual field measurement was repeated up to 3 times by each examiner, and the mean of each examiner's result was recorded. If there was a discrepancy of ≥10 degrees between the 2 examiners' measurements, both examiners performed the field test until a consensus was reached. Hemifield asymmetry of ≥20 degrees was considered abnormal. In addition, a decrease in hemifield values of ≥20 degrees from established normal patients also was considered abnormal.

Graphs were generated plotting the visual field data obtained from the patients with plagiocephaly against nor-

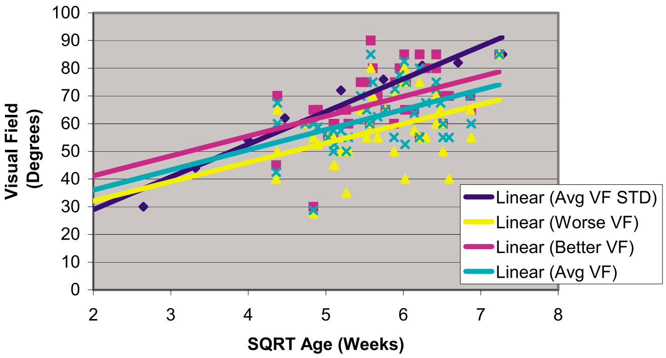

FIG 3. Graph of temporal visual darker field (degrees) versus square root transformation of age (weeks). Black, Normative visual field data; white, worse hemifield data; lighter black, better hemifield data; and gray, average visual field data.

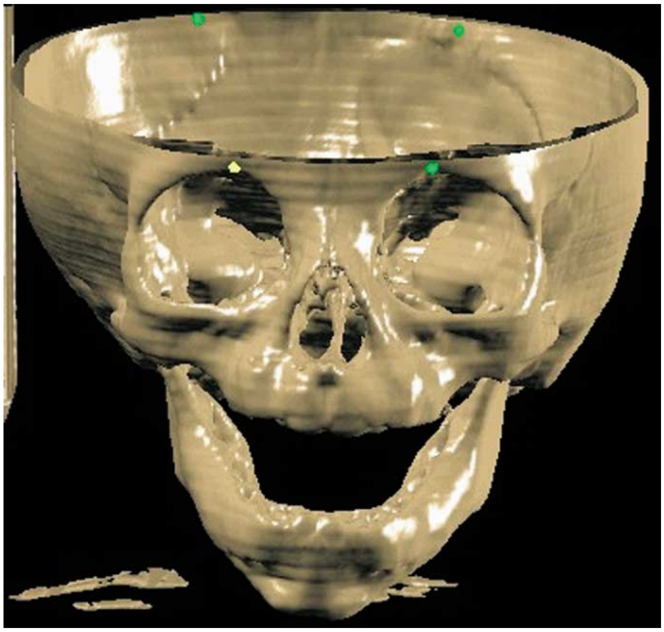

FIG 4. CT reconstructed image measuring the distance between the anterior supraorbital notch and the contralateral occipital bone.

mative data extracted from the study by Mohn and Van Hof-van Duin[14] (Figure 3). The square-root transformation of the age was graphed against visual field in degrees to establish a normative line for visual field development. We then compared the heights of the regression lines generated by the data obtained in our study to the normative line using a one-way analysis of covariance.

Three-dimensional computed tomography (CT) of the skull was performed in 16 of the study patients to obtain objective data on the severity of plagiocephaly. CT images were received from the radiology department on CD-ROM and transferred to the Panchal Imaging Lab, University of Oklahoma College of Medicine, Division of Plastic Surgery. The data were then copied to the hard drive of the imaging workstation (DELL Precision P-530 with dual 1.5 GHz processors, 500 MB RAM and 100 GB RAID 1 mirrored storage). The raw slices (2.5-mm thick) were imported in to Analyze 4.0 (Biomedical Imaging Resource, Mayo Clinic Rochester, MN) to be prepared for landmarking. The empty outer margins of the images were cropped to reduce rendered volume file size, and the volume was forced to cubic voxel dimensions. Proximal portions of the mandible and vertebral column were segmented out (when necessary) to allow visualization of exocranial base landmarks. Remaining artifacts of the CT scan gantry image were removed and the rendered volume saved in Analyze Image 7.5 format. The Analyze Image 7.5 volume was opened, and the bone threshold level for each image was determined empirically. A standard coordinate template file was opened, renamed, and saved under the patients' data archive with an identifier, date, and threshold level for future reference.

Collection of CT and Magnetic Resonance Images

CT Scans of the Head. Each CT scan was opened in exploratory 2-/3-dimensional image processing system (etdips) software. This imaging software allows the simultaneous visualization of the reconstructed volume in 3 orthogonal views, thereby allowing accurate placement of biologically relevant landmarks. Landmarks were chosen at the supraorbital notch anteriorly on each side. At the same CT slice level, the most prominent part on the occipital bone was chosen on the right and the left occipital bone (Figure 4). The right oblique diameter measured the distance from the right supraorbital notch to the contralateral most prominent point on the occipital lobe as described. The left oblique diameter was measured vice versa, ie, from the left supraorbital notch to the most prominent point on the right occiput. The absolute difference between the right and left oblique diameter gave the extent of asymmetry caused by plagiocephaly. Graphs were generated comparing visual field defect and laterality to cranial asymmetry to determine if a correlation existed between visual field defect and cranial asymmetry.

RESULTS

Visual Field Constriction

Constriction of one or both hemifields by at least 20 degrees compared with established normal patients occurred in 35% (14/40) of patients. Four (10%) patients (4/40) had constriction of both hemifields whereas 10 (25%) patients (10/40) had constriction of one hemifield. There was a significant difference between the worse hemifield measured (white line) in each patient and the standard visual field calculated from the normative data (darker black line) ($P = 0.004$). (Figure 3)

Hemifield Findings

Hemifield asymmetry of 20 degrees or more (intrapatient right vs. left hemifield difference) was found in 17.5%

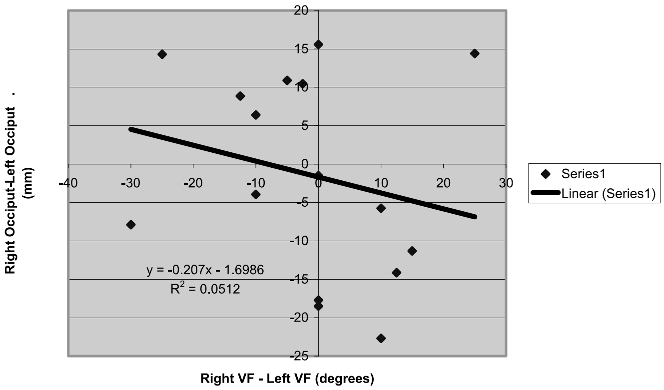

FIG 5. Skull measurement asymmetry (mm) versus visual field asymmetry (degrees).

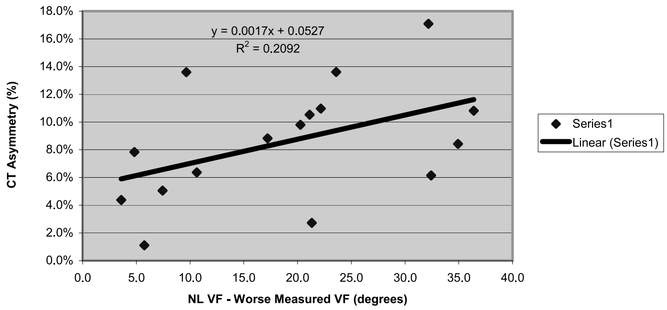

FIG 6. Degree of CT asymmetry (%) versus amount of visual field constriction (degrees).

(7/40) infants tested, with a range of 20 to 30 degrees. A similar plot of the better hemifield was not statistically different from the normative curve (P = 0.390) (Figure 3, lighter black line). A plot of the mean of the 2 hemifield measurements for each patient versus age was statistically different from standard visual field (P = 0.036; Figure 3, darker black line).

Visual Field Development

All data from the study population showed a seemingly delayed progression of visual field compared with the standard curve, ie, the slopes of all cohort data curves were shallower than the slope of the standard data group. Although this difference was not statistically significant (P = 0.147, analysis of covariance), it may indicate a trend of delay in visual field maturation in patients with posterior plagiocephaly.

Visual Field Defects Versus Severity or Laterality of Deformity

No correlation between laterality of visual field defect and laterality of plagiocephaly could be made on the basis of objective data obtained from 3D-CT (Figure 5). CTs from 3 of 16 patients showed visual field defects contralateral to the flattened side whereas 9 of 16 showed ipsilateral field defects. The remaining 4 patients demonstrated no visual field defects. A correlation between increased constriction of the worse hemifield from normal visual field for age and increased asymmetry on CT was noted (Figure 6).

Developmental Delay

On the basis of parental responses, 12% (5/40) children had some type of developmental delay or did not appropriately meet expected milestones for age. Formal data from pediatricians or neurologists were not available in this regard.

DISCUSSION

Risk factors for deformational posterior plagiocephaly include male sex, first-born, prematurity, and sleeping in the supine position only. These children often are perceived to be less active, have developmental delay, and a preferred head orientation by 6 weeks of age.[9] Localized cranial flattening (which occurs in 13% of healthy newborns) at birth may predispose to deformational posterior plagiocephaly.[6] Mulliken et al[3] proposed that the fetal head becomes distorted as the result of passage through the maternal bony pelvis, which would account for the higher incidence of right parietal and left anterior plagiocephaly as expected with the left occipital anterior passage through the birth canal.

A retrospective review of 100 patients with positional posterior plagiocephaly found that 64% had sternocleidomastoid imbalance in muscle mass, muscle strength, or both, and that 12% had torticollis.[16] Such sternocleidomastoid dysfunction or weakness may also predispose children to deformational posterior plagiocephaly. As a result, these children develop flattening of the parieto-occipital area and an anteriorly shifted ear contralateral to the shortened sternocleidomastoid or ipsilateral to the weak sternocleidomastoid. The patients with sternocleidomastoid imbalance also frequently have an intermittent head tilt and favor rotating the head to one side. In patients with sternocleidomastoid imbalance caused by weakness, the prone positioning may actually encourage strengthening and stretching of neck muscles through frequent head lifting.[16] Alternatively, a child could conceivably develop sternocleidomastoid contracture from nonmuscular plagiocephaly, although this process may take weeks or months to occur.

Although it was previously thought that posterior plagiocephaly posed only cosmetic problems, Miller and Clarren[17] showed that 39.7% of patients with deformational posterior plagiocephaly required an Individual Education Plan, with services such as special education, speech therapy, physical therapy, and occupation therapy. These results show that infants with deformational posterior plagiocephaly are at a higher risk for future developmental difficulties, with subtle problems of cerebral dysfunction such as learning disabilities, language disorders, visual–perceptual problems, motor delays, and problems with attention span. At the initial evaluation of these patients, 13% had a history of concerns of developmental delay. Panchal et al[18] evaluated the incidence of neurodevelopmental delay in children with deformational posterior plagiocephaly and found that, depending on the test used, 20-48% of children had mild developmental delay and 9-13% had severe developmental delay.

Few longitudinal studies exist that evaluate the prevalence of positional preference in children. A study of 623 infants in the Netherlands showed a prevalence of 8.2% at 16 weeks of age. At the first follow-up study at 7-14 months, 47% had asymmetric flattening of the occiput, decreasing only to 45% at 2 to 3 years of life.[7]

This study demonstrates that deformational posterior plagiocephaly can affect visual development in a quantifiable manner. On the basis of our study, there is a notable incidence of visual field constriction in patients with deformational posterior plagiocephaly. Another hypothesis is that patients with deformational posterior plagiocephaly may have delayed progression of visual field development, whether attributable to the plagiocephaly or to some global developmental delay. This study also demonstrates that posterior plagiocephaly can be quantifiably measured but neither the laterality nor the severity of skull deformity is predictive of the severity of visual field defects.

The expected result of unilateral flattening would be a contralateral hemifield defect. Interestingly, the laterality of the skull flattening and the laterality of the visual field defects were not predictive of one another. We are unable to explain this finding. Invoking the "coup contre coup" forces that occur with head trauma as playing a role in this condition seems unlikely and difficult to conceptualize.

Limitations of this study include those inherent in performing visual field testing in infants because of cooperation. Also, we relied on established normative data from the literature rather than repeating these studies in our clinic. Additionally, there is a possible referral bias, with only the more severe cases of deformational posterior plagiocephaly being sent for evaluation at a major academic center. Further studies are needed to determine the natural history of visual field development in posterior plagiocephaly, as well as to evaluate any effect helmet therapy may have in this regard. It is possible that spontaneous normalization occurs in most cases; such improvement in gross motor milestones in children who sleep supine has been seen by 18 months of life.[19] Nevertheless, our data lend credence to the concept that posterior plagiocephaly may be associated with clinically measurable neurologic sequelae. These findings may be due to direct effects of the plagiocephaly itself, or related to a more global developmental delay.

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